**PODCAST 39: Cystic Fibrosis-Related Diabetes**

**Dr. Rita Kalyani, MD:** Welcome to Diabetes Deconstructed, a podcast for people interested in learning more about diabetes. I'm your host, Dr. Rita Kalyani at Johns Hopkins. We developed this podcast as a companion to our patient guide to diabetes website. If you want a trusted and easy to understand resource for diabetes or to listen to previous podcasts, please visit hopkinsdiabetesinfo.org.

For today's podcast, it is my pleasure to introduce a person with diabetes, Joanna, who will be sharing her journey, not only living with diabetes but also her journey focused on her experience living with cystic fibrosis (CF), along with diabetes. Along with Joanna, we are pleased to welcome Dr. Aniket Sidhaye, an expert on cystic fibrosis-related diabetes (CFRD).

Dr. Sidhaye is an associate professor of medicine and pediatrics at the Johns Hopkins University School of Medicine. His specific clinical interests are cystic fibrosis-related endocrine disorders, type one diabetes, particularly emerging in emerging adults with diabetes, thyroid disorders, metabolic bone disease, and management of hospitalized persons with diabetes.

Dr. Sidhaye earns his MD from Northwestern University's Feinberg School of Medicine. He completed his residency at Northwestern University as well. He performed a research fellowship in endocrinology and metabolism at Northwestern University and also performed fellowships in endocrinology and metabolism at Johns Hopkins. In addition, he is a committed educator serving as an advisor in the medical school, co-director for the endocrine course for preclinical medical students, and program director for our endocrine fellowship. Welcome, Joanna, and Dr. Sidhaye.

**Joanna,** a person with CFRD: Thank you.

**Dr. Aniket Sidhaye, MD**: Thank you, Dr. Kalyani, for inviting us to do this, and especially thank you to Joanna for spending time with us today. I thought I would begin just by explaining what cystic fibrosis is—cystic fibrosis is a genetic condition that affects about 35,000 people here in the United States of America. It is a condition that is caused by a defect in a molecule that's very important for maintaining water and electrolyte balance in the body, and it affects many different parts of the body, but in particular, it affects the lungs, and that is what causes the most problems for patients. But it also affects the pancreas.

Interestingly, it affects the part of the pancreas that is responsible for helping us digest foods properly; people often have malabsorption. But as part of the damage that is occurring to the pancreas, the part of the pancreas that's responsible for making insulin is also damaged. So it turns out that people with cystic fibrosis are at a higher risk for developing diabetes.

In fact, for people with cystic fibrosis above the age of 30, about a quarter of them are affected by diabetes. In addition to a lot of the burden of taking care of cystic fibrosis, which you're going to hear about from Joanna, diabetes is also something that many patients have to contend with.

So I thought here it would be good to turn it over to you, Joanna, and hear about your journey with diabetes and with CF.

**J:** Yeah, thanks for having me. I was born with CF. When I was first born, my parents had never heard of cystic fibrosis. So, they didn't really know much about what was going on. I was born in 1976, and back then they didn't screen newborns at that time for cystic fibrosis. And my mom knew that there was something wrong, but she wasn't really sure what. I was her first child. She was a new mom, just not really sure what to do [or] what was normal for a newborn. My mom had taken me to the pediatrician, and the pediatrician said, “I think she has asthma, and she probably just has a lot of food allergies, and that's why she's not gaining weight.” That was what my parents were told. Then my sister was born 13 months after me. My sister Noelle was born with a lot of complications at the time, and they tested her for cystic fibrosis. She was actually diagnosed before I was, and then they diagnosed me. And my parents knew that Joanna had CF. So that's how I found out I had CF—I was one when I was diagnosed. I have two other brothers and sisters who don't have CF, and we lived a pretty normal life.

My parents tried to get us more physically active. That was a big thing, trying to get us to do more for our lung function, so we always were trying to do sports and things like that, especially in high school and in college.
My sister had a lot more complications than I did, and she did pass away from CF in 1996. She was waiting for a lung transplant. It's a very devastating disease, and it's very taxing. My sister was diagnosed with diabetes before I was. I knew that the classic symptoms when she had it—she was very thirsty and hungry, and they tested her, and she had diabetes.

Then it wasn't until a couple years [later] I was in college when I got diagnosed. I think there was some time that they weren't sure. I don't know because I didn't see what my glucose numbers were, but I knew I had tendencies, but I wasn't really told I had diabetes for a long time. I went through stages where—if they think it's not a big deal, I'm not going to think it's a big deal. I remember right before I went off to college, they told me, “Just watch what you eat.” That was it, that was all they told me. They just said, “Watch what you eat.” I did have a glucose meter in college. I remember every time I tested my sugar, it was high. But I don't know; they don't really think it's a big deal. It wasn't until I was home for one summer and I was taking a summer class, and I was extremely hungry. I was having blurry vision, and I had just had a CF tune-up—that's what we call when you go into the hospital for IV antibiotics. I would go in every year just to pump myself up for my lung function. And I remember I went in, and I was seeing a new pulmonologist. I had just had this tune-up, and I went in for a visit, and he said, "So how are you feeling?" And I said, "I don't know. I don't feel great, but I don't know what's wrong." He says, “Let's go get some blood work done today." He made me wait around for the blood work. And he said, "Your blood sugar is in the three hundreds, and your A1C is 12." So he made me go to an endocrinologist that day.

But before this, I did have tendencies. They did try to put me on a sulfonylurea, and I didn't do really well with it, [so] I just stopped taking it. And I thought—I have a new doctor; he doesn't know that this is a problem for me; I'm just hoping it goes away. So that's how I started with the diabetes, with the CF

**AS:** Yeah. Thanks for sharing. I think your story highlights so many things that are challenging. First and foremost, CF is a condition where there have been really remarkable changes. Where 50 years ago, most people with CF did not live into adulthood. When I was in training or when Dr. Kalyani was in training, people had started to live into their early twenties. And now in North America, the average lifespan is just above 50 years of age. There have been transformative changes. But I think many years ago it was not so well recognized how common cystic fibrosis-related diabetes is.

But when initial studies started to be done, people realized that right before people get diagnosed, there's actually a drop off in lung function and people's weight goes down. They start to feel and do more poorly, and diabetes is a risk factor for bad outcomes in people with CF. Because of that, now there are screening programs where everybody above the age of 10 who has CF is supposed to get a screening test for diabetes.

Usually that consists of something called an oral glucose tolerance test. You mentioned the hemoglobin A1C; yours was very high. Probably many people listening to this podcast will know that an A1C above 6.5 percent constitutes a diagnosis of diabetes, but [for] many people with CF early on, their A1Cs are not that abnormal. And I wonder if maybe that was the case with you, but if they had done the oral glucose tolerance test, they would have seen just how much abnormal glucose there were.

I think the other thing is [that] we think of cystic fibrosis-related diabetes as a condition that really requires insulin to be treated because of the damage to the pancreas. The amount of insulin production is really very low. And so medicines like you described—sulfonylurea, which is a medication that tries to get the pancreas to make more insulin on its own—end up being not so effective. I think nowadays, based on publications and guidelines and research, most people start with insulin because they know that helps people. I would be interested to know once you did get started on insulin, or if you got started on insulin right away, how you did feel.

**J:** I did. I started on insulin that day, and I remember they made me come back the next day to talk a little bit more about how to dose the insulin, and we went over diet information. I remember at the time I felt like it was a manageable part of my health. I felt like it was a game. I know that sounds really weird, but I thought, "Oh, let's see if I pair my insulin with what I'm eating. Let's see if I did a good job."

And for me, it was fun. I thought it was really fun. I don't know if I'm normal or not, but for me, I thought it was really neat how I could take my insulin and I could bring my sugar down based on what I was eating. So, for me, I thought that was really a neat thing because with CF, you don't always have control. I felt like with diabetes, there was a little bit more control that I could make an impact with my blood sugars.

**AS:** Yeah, that's an interesting perspective. Many people with CF that I've spoken with find the diagnosis of diabetes just to be such a burden because you're already describing having to dose insulin with every meal that you eat. I wonder for the audience; it might be good for them to hear about what it took at that time. I know there've been lots of changes more recently, but at that time when you were diagnosed, what did you have to do to take care of your CF? Because taking care of the diabetes was now on top of that.

**J:** I was in college at the time, so that is a hard time to get diagnosed with diabetes. Plus, I had CF. I did the best I could. I went to a small college where I was playing sports. So, I didn't go to a big school where maybe I couldn't play a sport, but I went to a school where I was playing field hockey. It forced me to run and do more physical activity, and that I think really helped me. I think from a lung perspective, it helped me stay healthy throughout the year. So, when I was away from home, I feel like the physical activity helped me with that.

I did have to do chest PT a couple times a day. I had a vest that I took with me to college, and I didn't really do nebulizers at the time, but I took pancreatic enzymes, and I had to take my insulin, and I was on NPH (Neutral Protamine Hagedorn) insulin and Lizpro; that's what I was on at the time.

But I felt for me, the CF was there, and as long as I just did my tune-ups, and I did my therapy. I'll be honest with you; I was a typical college kid; I didn't always do my therapy all the time. But I was lucky that I didn't have a problem with that, and it wasn't until I got older that the CF lung disease became more of a problem for me as I got older, but when I was younger, I could get away with it a little bit because I think I was more physically active.

**AS:** Yeah, no physical activities—very important for CF and for diabetes, it turns out. But I think it is important to note that many people have to do chest physical therapy, which involves not a small amount of time, multiple times a day. They might have to do certain types of nebulizer treatments where they're taking in a medication and inhaling a medication again, multiple times a day. It sounds like you and maybe your family have tried to make the best of the situation so you could also handle the diabetes, but to layer on taking multiple injections of insulin is indeed quite a lot of work. For all the people that do it, they're really to be commended because it is something. that, as providers, we have to be cognizant of the mental health aspect that goes along with that. How did you feel after taking insulin? Did you notice if it made you feel any better?

**J:** I did feel better: my vision got better, and I wasn't as tired. I definitely feel like it was helping me with my energy level, and I felt better. When I graduated from college, when I first started working, that was really tough too, because [of] the demands of my health and also working—I wasn't used to that. And I didn't have my mom and dad looking over my shoulder so much. But, I just remember, as diabetes has progressed over the years, just simple things like using an insulin pen was so much easier than using a vial. I remember talking to my endocrinologist, and I'm like, "It's such a pain in the neck, these vials." We have pens now; that was easier. And then, years later, I did go on to an insulin pump as well. There

are some things I think with the diabetes that have made my life easier to manage. It is another burden with my health, but I do think that there have been some things that have helped people live a better life with diabetes.

**RK:** Joanna, I really appreciate you sharing your story about the interplay between the CF and the lung health and the diabetes, which is related to it. It's interesting—we talk a lot about type one and type two diabetes. I think for many people listening, perhaps they're not familiar with the idea that you can have diabetes as a result of another condition for [example], in your case, diabetes related to the cystic fibrosis. I'm curious; what did your doctors tell you? Did they have a hard time defining the kind of diabetes you had, and what did you tell people when you had to share your diagnosis? Were they trying to categorize you into type one or type two, or did you find that you have to explain the kind of diabetes you have?

**J:** I always knew that it was not type one; it wasn't type two; it was somewhere in the middle. And I struggled with that too, a little bit like—I take insulin, but I'm not type one; it's not autoimmune, so I did struggle with that. I had some friends that didn't quite understand what type of diabetes I had; they knew I had diabetes. I remember one time I was on the van when we were going to a game when I was in college, and I was having low blood sugars, and one of my friends said, "Give her insulin!" and I'm like, "No, don't give me my insulin now! I don't need my insulin right now." So I think my friends were very concerned because they didn't quite understand what type of diabetes I had, but they knew I had CF, and they knew there was a lot to that. I think I struggled with that. I don't know if my doctors really gave me a good explanation of really what was causing it, and I struggled with that a little bit too. Why do I have diabetes? I knew my pancreas was involved, and I didn't always know—was it because my pancreas just wasn't working? Was it scarring? What was the problem? I didn't really understand it.

**AS:** What's interesting from a historical perspective is [that] although we associate cystic fibrosis with a lung disease, and indeed that's the reason that most patients really suffer—the lung disease. Cystic fibrosis actually refers to cystic fibrosis of the pancreas. And so, the person who really discovered cystic fibrosis was a woman named Dorothy Anderson, who was a pathologist at Columbia at the time, and I only mentioned her because she was a graduate of the Johns Hopkins University School of Medicine, [and] this podcast is hosted at Hopkins. But it really was these cysts that formed in the pancreas, and then the scarring that occurs in the pancreas that then led to damage to the parts of the pancreas that make insulin. And in a way, not exactly, but something similar is happening in the lungs because of the inflammation in the lungs. And with the repeated infections that many people get, there starts to be scarring in the lungs and damage to the lungs. So one of the things we hope when we give people an anabolic hormone like insulin, especially in people with cystic fibrosis, is that they pick up weight and maybe their lungs might get a little bit better. Did you notice if anything like that happened?

**J:** I did gain a little bit of weight with it. I don't know; my lung function was pretty stable at the time. So I didn't really notice a difference with my lung function when I was diagnosed. But I did start to put the connection together that when I was sick, if I kept better control of my sugars, I got over colds quicker and I got over the infection better or faster. So for me, that was something that I was really zoning in on and a motivator to try to keep my sugars under better control. Because if I did that, I felt better because, if I was sick, I felt lousy anyway. Then, if I could just control my sugars and get over this infection a little bit better, it helped me. I put that connection together, and it helped me to stay motivated to take care of my sugars.

**AS:** Thank you for sharing that, because not just in people with cystic fibrosis, but even in people who have type 1 or type 2 diabetes, there's an increased risk for respiratory infections, meaning [infections] of the lungs. Better glucose control is associated with not having as many of those infections. And of course, people with cystic fibrosis are at risk for many more infections of the lungs. It's really great that you put that together and took it as a motivator.

**J:** Another thing with that is if they had to use steroids—it was a nightmare. It is a nightmare. If, you know, I have to go on steroids, it's a nightmare to try to get it under control.

**AS:** Steroids, so medications like prednisone that people might have heard of, make the body more resistant to the effects of insulin. So even though you already take insulin, that insulin just doesn't work as well anymore. In particular, the insulin that you take around mealtimes does not work as well. I don't know if you noticed that your basal amount of insulin maybe didn't change too much, but the amount of insulin you had to give yourself at meals just really goes up dramatically. And in particular, in people with CF, it goes up more.

**J:** I did notice that.

**AS:** One of the things I did want to talk about was the remarkable changes that have occurred in the treatment of cystic fibrosis. And maybe I'll give you a chance to talk about some of these newer treatments that have come online in the last, five to seven years and how they've affected your lung function.

**J:** I have definitely benefited from some of the new drugs that are available. The new modulators that are available, and I'll let you talk a little bit more about that.

I was lucky enough right before COVID to be able to take Trikafta, and that was such a game changer for me. So, I felt better almost immediately. I would say within two days, I started feeling better. But I actually did have a problem when I first started taking it. It was almost like a shock to my system, and I actually had to go into the hospital. I had some GI issues with it, and I actually was throwing up and things. Then once we fixed that, I did okay with it.

But I knew that they were worried that maybe I wasn't tolerating it, but I was really hopeful. I knew that modulators were something that was coming out, and there were earlier ones that came out before Trikafta. And I was desperately trying to get onto a research study because I was really having a hard time with my CF. I was doing my chest PT, like at least twice a day, and I was doing eight nebulizers a day just to stay afloat. So I actually had to stop working because all I could do was really just take care of myself. So I was just doing basically my chest PT, my nebulizers, and just trying to take care of myself around, maybe 4 or 5 years ago. Then, when I was able to take Trikafta, I started feeling better. My lung function went up like 20%. My insulin needs actually went down.

**AS:** Oh, that's interesting.

**J:** Yeah, it was really cool. I remember telling my pulmonologist, "I feel like I'm on vacation, this is what it's like to be able to breathe!" It was just such a game changer. I was so thankful that happened before COVID. And I know that there [are] some people who can't take Trikafta or the other modulators. But I think it's made people with CF be healthier.

Obviously, I feel like I have a future and that's actually why I actually went back to work because my husband said, "Listen, Joanne, I think you can go back to work now."

**AS:** Yeah, I'm an endocrinologist, and mostly it's pulmonologists that really direct the care of people with CF. And so it has really been, just a privilege to be on the sidelines watching what these newer treatments have done, and it is a true triumph of modern medicine.

First off, all the other care things that have been done for people with CF in terms of the antibiotics and how they're administered have already helped people to live longer and more productive lives. I mentioned before that CF is due to a change in the molecule that helps the body transport chloride and bicarbonate; those are two of the electrolytes. And there are several different types of things that can occur, but either that molecule can't get up to the membrane of the cell where it needs to do its job or once it gets there, it doesn't do its job properly. So these newer modulators, as you call them, are just those small molecules that either help the molecule transport chloride and bicarbonate better or it helps the molecule get from inside the cell up to the surface better where it can do its job.

So the one that you mentioned, Trikafta, is a combination of three different pills that helps this molecule do its job better. And I've heard the same thing from other people. They say literally the next day or two days later, they stopped coughing, and it is just a remarkable thing to hear. And with regards to diabetes, as endocrinologists, we are all anxious to see if people with CF who also have diabetes—whether these modulators are going to help their diabetes get better.

I think the jury is still out, and we're going to have to see how it goes. I think for some people, they describe needing less insulin. For some people, they are able to gain weight and eat better, and they actually needed more insulin in the initial stages, right after they started Trikafta, and so there was a lot of adjustment on both ends with regards to diabetes. Now these modulators are approved for use down to the age of two. So what's really going to be important for the research community is what is going to happen to the risk of diabetes or for younger people as they get older? Hopefully, it will be less, but yeah, it's interesting to hear that you need a little bit less insulin. I guess you had to be vigilant for low glucose when you started Trikafta.

**J:** When I first started, yes. And then I did gain a lot of weight because I was so used to a CF diet, which is eat as much as you want. I watched my carbs, but I was never calorie restricted. I had to look at what I was eating a little bit better to get my weight down a little bit because I did gain some weight.

**AS:** That's actually another great point about how cystic fibrosis-related diabetes was, and maybe still is, different than type 1 or type 2 diabetes, and maybe you could just talk about that a little bit because people with cystic fibrosis tend to not absorb calories very well, and they're underweight, and that weight really contributes to their lung function.

In CF care centers, it's a big priority to help people get their weight up. Sometimes that is at odds with managing diabetes. Could you talk about that a little bit?

**J:** That is something like, growing up, I could eat whatever I wanted; no one ever said anything. I actually thought it was funny that I could have a high-fat diet. This is my diet. It was hard to go when I started gaining weight when I was on Trikafta. I was like, "Oh, wait a minute." Now I'm a normal person who really does have to watch their calories and things like that. My friends used to say, "Yeah, now you're normal. Now, you know what it's like."

It was a swing from one direction to another as far as diet goes. And even when I was in the hospital, I remember before Trikafta, the endocrinologist would come in to visit me, and she would get very upset about my diet because it was a high-fat, high-protein diet, which is what they would order for me. She would come in, and she'd say, "What are you doing eating all this food?!" So, I felt like every time I was in the hospital, my diet order would change multiple times, from a diabetic diet to a high-fat, high-protein, double-portion diet. So, it was something; it was totally different. I'll be honest with you, what Trikafta has done for me and my life is totally worth it.

It's so nice to have my life back and to be able to breathe, and I feel like I'm going to live longer than I ever thought I would. So, it's really amazing times to have CF at this time. Because I know what it was like before and I know what it's like now.

**AS:** One thing that is true is that even before this one specific modulator came out, there were other modulators that had come out before that. So Trikafta is a combination of Elexacaftor, Tezacaftor, and Ivacaftor. And so ivacaftor had come out before, and it can be used for people with CF who have very specific types of mutations.

And there are other modulators that came out in between as well that also had good effects, but we're now on to one that seems to have an even better effect, and you mentioned one thing: there's about 90-95 percent of people who are eligible for these modulator types of treatments. But there are patients that aren't eligible for these modulator treatments because of the type of CFTR they have. I know that it's an active area of research. One thing about cystic fibrosis is what an amazing, I think, effect many of the foundations that advocate for patients with CF and for research in CF; they've had a huge effect on really pushing the science, pushing the clinical care. And so now we have these; they're called highly effective modulator treatments, but now they're pushing for treatments so that all patients with CF will have a meaningful treatment and so that they can also have these types of benefits.

**RK:** That's so neat to hear about all these new treatments and it sounds like a really exciting time, to have these options at a minimum for people with CF.

Dr. Sidhaye, I wonder if you could talk a little bit about prevention of diabetes in people with CF. Joanna, you mentioned the weight challenges and how with CF, you're encouraged to gain weight, high-calories, high-fat, yet in the context of prevention of diabetes, those are perhaps the exact opposite of what we want. Is there research being done into prevention of diabetes in CF?

**AS:** That's a good question. I think that this is so fundamental to why CF-related diabetes is different from type 1 and type 2 diabetes. In the pre-modulator era, I would say that getting a person with CF and making sure their weight is where it needs to be to help preserve their lung function was a priority. And so we would have to balance. Certainly, advise some restriction of calories and carbohydrates, but not anywhere to the degree that we would for people with type one or type two. And that's because gaining weight was so important. Insulin is the way that we make that happen. But now we're in a different era, and modulators mean that, if I remember the numbers correctly, just about a third of people with CF can be classified as overweight or obese, which is a totally different place than it was 10-15 years ago. I think we are going to have to start thinking about CF-related diabetes a little bit like how we think of type 2 diabetes.

We are going to have to start thinking about that and counseling patients in ways that we never did. I know that we have even started using medicines that we might use in type 2 diabetes for people with CF. At this point, it's really just single cases done by individual practitioners, but there are studies ongoing about this.

I definitely think that in the modulator era, CF care centers and providers who care for persons with CF are going to have to figure out how to counsel people about risk factors for diabetes. I definitely think that if there's a family history of type 2 diabetes, then that is something that we all should think about when we meet people with CF who might be at risk for diabetes and start counseling them about their weight. But this is not a conversation that anybody would have even thought of having just five or six years ago. I think it's not even on the cards.

**J:** It's funny you say that because I went on an insulin pump. I thought, number one, it would make my life easier in some ways, being on a pump, because it was a little bit more convenient to take my insulin. But I felt because my energy needs were so high and my calories were so high, it was easier for me to control based on having a pump. But now I have to watch what I weigh and what I eat, but I still have a pump, which has helped big time. But I use it a little differently and realize it's not a free for all, and I still have to watch what I eat.

**AS:** I'm glad that this conversation has drawn parallels. There have been remarkable changes in the care of CF. But there have also been remarkable changes in the care of diabetes that you yourself have just described from being on NPH and regular insulin with vials to pens and now insulin pumps. And I'm sure you use a continuous glucose monitor as well.

**J:** Yeah.

**AS:** All of those things have made the control of diabetes, I want to say in some ways easier. I know that a lot of work still has to be done by patients.

**J:** It's definitely a lot easier than what it was, and CF has gotten a lot easier than what it was because of these different drugs. I feel like I'm living in a really interesting time to be able to see, and I don't know if people, in the future who have CF, are going to really understand what it was like for people who didn't have these drugs or even the technology with diabetes.

**RK:** Joanna, I found it fascinating when you talked about your perspective about feeling like you had more control when you were diagnosed with diabetes compared to the diagnosis of CF, which was still emerging. There was a lot still not known at the time you were diagnosed. While it can be a burdensome disease, it was interesting for me to hear your contrast and how you felt like you had a say in what was happening [with your diabetes] and in some ways made it a fun game in order for you to be able to manage effectively. I wonder if you could talk a little bit about that empowerment that you felt, the fact that you had a role in the management of your disease.

**J:** I don't know how it happened. I do feel like with CF, there are so many variables that you don't have control over. I just put the connection that I can really control my diabetes. And I know a lot of people with CF; I know friends with CF; I know it's not [easy]; it's a hard thing because it's one more thing you have to worry about—your blood sugars. But for me, I felt it did help me control my health, and I felt like it did impact my CF. So, for me, it would seem natural to me to do that. But I know I've been lucky with CF; I really have. I know people like my sister who weren't very lucky with CF. You can get an infection, and it can really take over your life, and you have no control over it. I've gone through periods where I didn't think I was going to get better. Before I had these new drugs, I wasn't sure if I was going to get better. And I think I was heading the road towards a lung transplant. I think I was on my way. So, I think with having the CF and the diabetes, it was just one aspect that I felt like I could control.

**AS:** Think it's a good message even for us providers. That maybe for certain patients, couching it in those terms may be a motivating factor.

**J:** Yeah

**RK:** And clearly the healthcare providers you saw, I'm sure, had a big impact on how you were able to manage your disease. And Dr. Sidhaye is an expert in the field of cystic fibrosis-related diabetes, but I imagine not all the providers you saw were as familiar with the management of diabetes. How do you think you were able to learn more about how to manage this particular type of diabetes?

**J:** I always took it on myself that I felt like I was in charge. I tried to be in charge. And I knew that a lot of the doctors I saw for diabetes might not know anything about CFRD. I'm probably the only patient they've ever had with this. I don't want them to treat me like a type 1. I don't want them to treat me like a type 2. But I took it with a grain of salt, and I felt I [could] feel my way out and do what's best for me with everyone. I think there's aspects of treating type one and there's aspects of treating type two that I can use for my own health. So even though maybe they don't know anyone quite like me, I think I can try to make this work for me the best way. And I don't know if that makes any sense, but I tried to do that.

**AS:** I know that there's a push by, in North America at least, the Cystic Fibrosis Foundation to actually educate endocrinologists in the care of persons with cystic fibrosis. That's the way I got involved and learned more about it myself. And so I think the major professional organizations, not just for CF but probably in many other situations, do have a role to play in making sure that we get educated, and it's on us to get educated as well.

**J:** I think that's great because people with CF [may] feel like you're not in one basket; it's not a cookie cutter. And I think sometimes you have to figure out—all right, it's nice that they have people that are being trained to understand the CF-related diabetes because it isn't type one, it isn't type two; it's a little bit different.

**AS:** And I think we have to be sensitive to that.

**RK:** Dr. Sidhaye, I wonder if you could talk a little bit about the complications of diabetes. Are they the same in people with CF who have diabetes versus not? And Joanna, I wonder if you could talk about what routine screening you undergo for complications from diabetes as well.

**AS:** Yeah, that's a good question. The typical long term complications we think of: eye damage, kidney damage, and damage to the nerves, they can occur in people with cystic fibrosis related diabetes, and they do occur. It seems to be that they don't occur quite with the same frequency. That is to say there are some people with cystic fibrosis related diabetes whose fasting glucoses are actually pretty normal and it's only the post meal glucoses that are a problem. Those people seem not to be at such a high risk for these types of complications that I did mention. But otherwise, if they do have progression of their cystic fibrosis related diabetes, they are at risk for those complications.
Now, we always think of diabetes as being a risk factor for cardiovascular disease, so heart disease and stroke. I would say until very recently, that was almost unheard of in people with CF. And that's another place we're in a new era where we are going to have to start worrying about that and counseling people. But we don't know how to do it yet. For the other complications, we do screening, and I think Joanna could probably tell you if she does or how often she does it.

**J:** I do the regular screening like most people with diabetes. So I get my eyes checked once a year for a dilated eye exam. My endocrinologist checks my feet to make sure I have feeling in my feet. I get my blood pressure tested and cholesterol, which never really has been a problem. But I have noticed with these new meds sometimes I have issues with my blood pressure a little bit, which I never had. I guess with CF and just getting older in general, these are things I'm going to have to look at a little bit. They check my microalbumin once a year too. I just get the normal tests done that most people with diabetes get, and I get my A1C checked at least twice a year, every 3 months. So like everyone, if mine's at goal, they sometimes are a little bit more lenient with me about getting it checked, but I just try to keep up with it. And, the pump and

having a CGM has helped. I've lived a really normal life. I also had a baby. I've had a very normal life with just keeping on top of my CF and my diabetes, but I just keep checks and make sure all that preventive stuff is taken care of.

**AS:** I'm glad Dr. Kalyani brought that up because it is important that we do the normal screening as you mentioned for urine protein, for foot exams, and for eye exams in people who have cystic fibrosis-related diabetes. Because there are things that we can do to prevent those complications or at least delay them from occurring.

**RK:** Yes this has been such a wonderful discussion. It's provided such great insights into something that we're learning more and more about. But for people with cystic fibrosis, diabetes can occur at a higher frequency than in the general population. And it's important for people to be aware of this, especially if you have cystic fibrosis or even other medical conditions that put you at risk for diabetes. Dr. Sidhaye, thank you so much for offering your wonderful expertise. Joanna, I wonder if you might have some parting words for those individuals that are listening and who might be struggling to manage their diabetes.

You talked about some of the experiences you had, but what would you say to someone out there who, regardless of the type of diabetes, is struggling to manage it?

**J:** I would say, I think that the more you can learn about how to take care of yourself, it's going to help you manage it better, and all these things add up. I think, even just doing a little thing every day is going to make an impact on your health. I think all these little things can make a big difference in the end. So for anyone with CF or diabetes, I think it's important to just, do the best you can. I feel like with the diabetes part, with CF, there are some things you can do to improve your health. So that's what I would say.

**AS:** It was so great to hear from you. And we are so grateful for you giving your time and your perspective. And thank you Dr. Kalyani for inviting us.

**RK:** Thank you so much, Sidhaye and Joanna for being here and talking about this very important topic. I know I learned a lot and I'm sure that our listeners did too. So, thank you again.

**AS:** You're welcome. Thank you.

**RK:** I'm Dr. Rita Kalyani and you've been listening to Diabetes Deconstructed. We developed this podcast as a companion to our patient guide to diabetes website. Our vision is to provide a trusted and reliable resource based on the latest evidence that people affected by diabetes can use to live healthier lives.

For more information, visit hopkinsdiabetesinfo.org. We love to hear from our listeners. The email address is hopkinsdiabetesinfo@jhmi.edu. Thanks for listening. Be well and see you next time.